The Intercontinental Journal of Emergency Medicine

Case Report

DOI: 10.51271/ICJEM-0003

Lemmel syndrome: a rare cause of obstructive jaundice (case report)

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Cite this article: Hatipoğlu FS, İpekci A. Lemmel syndrome: a rare cause of obstructive jaundice (case report). *Intercont J Emerg Med*. 2023;1(1):9-11.

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Submit Date: 10/03/2023 **Accept Date:** 30/03/2023

ABSTRACT

Obstructive jaundice through periampullary duodenal diverticulum (PAD), without gallstones or neoplasms, was first described by Lemmel in 1934 as Lemmel syndrome. PAD can be diagnosed incidentally or suspected in patients with obstructive jaundice and nonspecific abdominal pain. It is a rare and benign condition that can be misdiagnosed as malignancy. Ignoring this, can cause morbidity and mortality. Definite diagnosis is made by Endoscopic Retrograde Cholangiopancreatography (ERCP). An accurate diagnosis is very important to ensure patient management and to avoid complications of delayed care. We present a case who coincidentally was diagnosed with PAD and thus obstructive jaundice.

Keywords: Lemmel sydrome, emergency medicine, obstructive jaundice, endoscopic retrograde cholangiopancreatography

INTRODUCTION

Abdominal pain is one of the chief complaints for presenting at emergency department. Although it is a reflex to associate the pathology with choledocholithiasis or pancreaticobiliary tumors in patients presenting with right upper quadrant pain and obstructive jaundice, a pathophysiology caused by periampullary diverticulum (PAD), which presents with similar signs and symptoms should be considered.[1] In rare occasions that PAD causes mechanical obstruction of the main bile duct from the distal section, it is termed as Lemmel Syndrome.[2] Lemmel syndrome is difficult to diagnose because it mimics other pancreaticobiliary diseases and malignancies.[3] It is an underdiagnosed and underreported syndrome due to the lack of specific pathognomonic symptoms or signs.[2] Duodenal diverticula are more common in adults aged 70's and older, and have a prevalence of 11%-22% in the general population in postmortem studies. It is independent of gender; however, the detection rate varies depending on the imaging modalities.[3,4]

In this case, we present a patient admitted to emergency department with right upper quadrant pain and jaundice. He was coincidentally diagnosed with PAD and developed obstructive jaundice as a result.

CASE

66-year-old male patient is admitted to emergency room with right upper quadrant pain that had been going on for a week. His known diagnoses are hypothyroidism, coronary artery disease, hypertension and hyperlipidemia. He has a history of gastric perforation, a horseshoe kidney operation and bilateral thyroidectomy.

On physical examination, patient's general condition was good, his consciousness was clear, his orientation was cooperative, Glasgow Coma Scale: 15/15. Vital signs were as follows: fever: 36.4, pulse: 79/min, blood pressure: 102/63 mmHg, sPO_2: 98% in room air, respiratory rate: 18/min. Abdomen was distended, no defense/rebound, rectal examination was normal stool smear. Last defecation was 2 days ago, he had flatus. Lung examination was normal, without rale/rhoncus. Posteroanterior chest x-ray was obtained with no pneumo/hemothorax (**Figure 1**). Cardiac auscultation was normal without murmur. Electrocardiography showed only mild incomplete right branch block.

Figure 1. The posteroanterior chest X-ray

Blood tests showed elevated liver enzymes and amylase. Hyperbilirubinemia with direct dominance was present (**Table 1**).

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Table 1. Summary of the Patient's Laboratory Values During the Hospital Stay

	Day 1	Day 2	Day 3		After ERCP Day 1	Discharge	Reference Range	Units
WBC	14.4	11.1	10		7.9	7.8	4.3 - 10.3	10^3/µL
HGB	13.3	11.9	12.5		11.2	11.3	13.6 - 17.2	g/dl
HCT	40.3	36.1	36.7		33.2	33.3	42 - 52	%
Creatinin	1.4	0.99	0.72		0.54	0.92	0.7 - 1.2	mg/dL
Urea	46	58	49		41	37	17 - 49	mg/dL
AST	382	199	447		139	71	< 40	IU/L
ALT	484	323	388		222	122	< 41	IU/L
ALP	637	492	622	ERCP	456	274	40 - 130	U/L
GGT	938	710	490		102	177	< 60	IU/L
Amylase	207	573	346		93	89	28 - 100	U/L
Total Bilirubin	7.85	6.24	6.91		2.44	1.08	0.2 - 1.2	mg/dL
Direct Bilirubin	7.16	5.81	6.66		1.92	1.05	< 0.3	mg/dL
INR	1.17	1.12	1.21		1.12	1.02	0.85 - 1.15	
C-reactive Protein	137	144	95		22	2.3	< 5	mg/L
pH	7.4	7.4	7.4		7.4	7.4	7.35 - 7.45	
Lactat	1.8	1.1	0.9		0.9	1.2	0.5 - 1.6	mmol/L

Imaging was conducted through hepatobiliary ultrasonography, no acute pathology in liver parenchyma was reported. Neither cystic or solid lesions nor perihepatic free fluid were detected. Gallbladder was enlarged, hydropic and 120×44 mm in size with 3 mm wall thickness. Sludge image that levels within the lumen was observed. Sonographically selectable calculus image was not detected in the lumen. No pericholecystic fluid was detected. Intrahepatic biliary tract appeared dilated and measured 4 mm at the distal site. Choledochus, pancreas and midline structures could not be evaluated due to the dense gas shadows. There was not any free fluid in abdominal compartments.

The patient underwent intravenous contrast-enhanced computed tomography (CT). Radiologist's verbal comment suggested a distended gallbladder, dilatation in intra and extra biliary tract, 1 cm diverticulum in duodenum's second segment that presses distally to the choledochus (**Figure 2**).

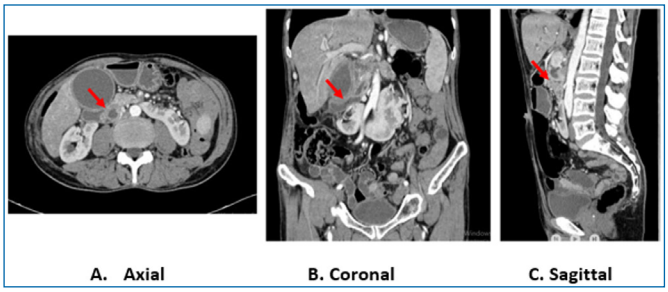

Figure 1. IV contrast-enhanced computed tomography (CT). The PADD is indicated by the red arrow at the A. Axial, B. Coronal, C. Sagital

The patient's oral regimen was stopped. Pantoprazole, metronidazole and saline infusion were administered. General surgery and Gastroenterology consultations were obtained. Patient underwent ERCP by gastroenterology that resulted in malpositioned papilla-sphincter oddi dysfunction, choledochal and intrahepatic biliary tract dilatation. Needle knife sphincterotomy (fistulotomy) and stent insertion into the choledochus duct were performed.

Patient was able to tolerate oral intake. His general condition rapidly improved. After final blood tests were obtained and showed regression in liver enzymes, the patient was discharged (**Table 1**).

DISCUSSION

Duodenum is the most common site of diverticula in digestive system after colon.[5] Duodenal diverticulum (DD) was first described by Chomel in 1710 and documented by Morgagni in 1762, and its first radiological demonstration was performed by JT Case in 1913.[5,6] PAD is an extraluminal diverticulum of duodenal wall located at a distance of 2 to 3 cm from the ampulla vater. Obstructive jaundice due to PAD, which is observed in the absence of gallstones or neoplasms, was first described by Lemmel in 1934 as Lemmel syndrome. Its incidence is 1 - 5% in radiological series, 11-22% in autopsy series.[1,7] The highest incidence is between the ages of 50 and 60, regardless of gender.[5,6,8] Although the physiopathology is not definite and specific, there are three main possible mechanisms:

1. Direct external compression of the common bile duct / ampulla, which leads to blockage,
2. Dysfunction of the sphincter of Oddi,
3. Irritation of ampulla, causing chronic inflammation and eventually leading to papilla fibrosis.[2,10]

A majority of diverticula are asymptomatic.[4,5,7] Even if only 1%-2% of cases may become symptomatic with acute abdominal pain in the form of biliopancreatic colic, our patient right upper quadrant pain that had been going on for a week. Leukocytosis, acute phase reactants increase, liver and/or pancreatic enzyme elevation, bilirubin elevation may be observed at patients' laboratory.[1,5,8] In our case, all laboratory parameters related to obstructive jaundice were elevated, so further examinations for definitive diagnosis were performed.

In addition, in 5% of cases, there may be various complications including:

- Non-pancreaticobiliary complications such as bleeding, diverticulitis, perforation or fistula formation.
- Pancreaticobiliary complications such as acute pancreatitis, cholangitis, bile duct stones or Lemmel Syndrome.

In the absence of choledocholithiasis or any other etiology of obstructive jaundice like in our case, Lemmel syndrome should be considered.[2]

Although ERCP is the gold standard diagnostic test; ultrasound, CT and Magnetic resonance cholangiopancreatography (MRCP) are first line diagnostic methods.

- Ultrasonography (USG) is usually preferred as the first imaging method in biliary obstruction since it shows the expansion of the bile ducts.
- MRCP is preferred when evaluating biliary tract anomalies and eliminating differential diagnosis to confirm the actual diagnosis.
- CT scan with intravenous and/or oral contrast is usually preferred due to its rapid acquisition and availability.
- In our case, we used a USG as a first imaging method, and it was non-diagnostic. Diagnosis of Lemmel syndrome was made by CT scan and confirmed by ERCP.

Since PAD is usually asymptomatic, treatment is only necessary when the patient becomes symptomatic. Its treatment in the literature is still controversial. Diverticulectomy is considered the gold standard treatment method. Nevertheless as a conservative medical treatment in oligosymptomatic patients, it is used in broad-spectrum antibiotic therapy in case of nasogastric decompression and perforation. In addition, endoscopic sphincterotomy or papillary balloon dilatation with ERCP is the first preferred treatment option because it is possible and has a good success rate. However, the recurrence rate is high in alternative treatments (the recurrence rate is 10%-24%). The cannulation rate in ERCP is 94.9%; complications such as bleeding or perforation are 7.8%. In case of unsuccessful endoscopic treatment, a surgical approach is preferred. But these procedures are difficult, their morbidity and mortality are high. Operative mortality is 20%-30%, while morbidity is 30%-40%. That's why perforation, etc. in cases without complications, the first choice is conceptive therapy or ERCP.[2,4-7] In our case, there was an Oddi sphincter defect and sphincteretomy was performed with ERCP.

CONCLUSION

Lemmel syndrome is a difficult diagnosis to establish and can be found by chance or suspected in patients with obstructive jaundice, pancreaticobiliary disease, nonspecific abdominal pain in the absence of stones or tumors like in our case. It is a rare and benign cause of obstructive jaundice. Differential diagnosis is important since it can mimic a malignant neoplasm. Ignoring this possibility, can lead to recurrent jaundice and, in some cases, cholangitis, which is related with high morbidity and mortality. Lemmel Syndrome should be considered in the etiology of obstructive jaundice and accurate diagnosis is essential to ensure proper patient care management and avoid complications of delayed management. Early diagnosis without complications as in our case, endoscopic or conservative approach may be preferred.[1,5,10]

ETHICAL DECLARATIONS

Informed Consent: All patients signed the free and informed consent form.

Referee Evaluation Process: Externally peer-reviewed.

Conflict of Interest Statement: The authors have no conflicts of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

Author Contributions: All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

REFERENCES

1. Bernshteyn M, Rao S, Sharma A, Masood U, Manocha D. Lemmel's Syndrome: Usual Presentation of an Unusual Diagnosis. *Cureus.* 2020;12(4):e7698. doi:10.7759/cureus.7698
2. Aourarh B, Tamzaourte M, Benhamdane A, et al. An Unusual Cause of Biliary Tract Obstruction: Lemmel Syndrome. *Clin Med Insights Case Rep.* 2021;14:11795476211063321. doi:10.1177/11795476211063321
3. Gao AR, Matta A, Seth R, Bande D. Lemmel's syndrome secondary to common bile duct compression by an inflamed duodenal diverticulum. *Cureus.* 2021;13(8):e16959. doi:10.7759/cureus.16959
4. Ergin A, Fersahoğlu MM, Kaya B, Onur E. Lemmel sendromu: duodenal divertiküle bağlı karın ağrısı ve sarılığın nadir bir sebebi. *Haydarpasa Numune Med J.* 2017;57(2):104–106. Doi: 10.14744/hnhj.2017.58077
5. Volpe A, Risi C, Erra M, Cioffi A, Casella V, Fenza G. Lemmel's syndrome due to giant periampullary diverticulum: report of a case. *Radiol Case Rep.* 2021;16(12):3783-3786. doi:10.1016/j.radcr.2021.08.068
6. Alzerwi NAN. Recurrent ascending cholangitis with acute pancreatitis and pancreatic atrophy caused by a juxtapapillary duodenal diverticulum: a case report and literature review. *Medicine (Baltimore).* 2020;99(27):e21111. doi:10.1097/MD.0000000000021111
7. Chavez JCM, Andrade LAM, Ramos ADJ, et al. Cholangitis secondary to Lemmel syndrome. case report. *Arch Clin Gastroenterol.* 2016;2(3):077-079.
8. Bellamlih H, Echchikhi M, El Farouki A, Moatassim Billah N, Nassar I. An unusual cause of obstructive jaundice: Lemmel's syndrome. *BJR Case Rep.* 2020;7(2):20200166. doi:10.1259/bjrcr.20200166
9. Azzam AZ, Alsinan TA, Alrebeh GA, Alhaider T, Alnaqaeb LJ, Amin TM. Lemmel syndrome as a rare cause of prolonged right hypochondrial pain: a case report. *Cureus.* 2021;13(12):e20093. doi:10.7759/cureus.20093
10. Oliveira DM, Correia C, Cunha F, Dias P. A rare cause of abdominal pain with fever. *BMJ Case Rep.* 2019;12(3):e228401. doi:10.1136/bcr-2018-228401